Case Report

Benign Retroperitoneal Cyst of Mullerian Type

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Retroperitoneal cysts are rare, usually asymptomatic, lesions. We present the case of a 42-year-old woman with an asymptomatic abdominal mass. The mass excised at our district hospital was found in the retroperitoneum. Histologically, the cyst was lined with benign Mullerian-type epithelium. The classification, aetiology and clinical manifestations of retroperitoneal cysts are discussed. Retroperitoneal cysts present as an acute abdomen in only a minority of cases. Ultrasound is the most reliable test for their detection, usually followed by computed tomography and/or an upper gastrointestinal series. Optimally, these cysts should be completely excised. [Asian J Surg 2004;27(4):333-5]

Introduction

Retroperitoneal cysts are rare lesions that, despite similarities in location and symptoms, are aetiologically and morphologically diverse. The incidence of such lesions is approximately 1 in 100,000 adult admissions. Mullerian cysts of the retroperitoneum are extremely rare disease entities and are considered to be a subtype of urogenital cysts. We report the clinicopathological findings in a rare case of benign retroperitoneal cyst of Mullerian type.

Case report

A 42-year-old female was found to have an asymptomatic right-sided abdominal mass on routine examination by her local medical officer. Her history was unremarkable, with no gastrointestinal, urinary or gynaecological symptoms. Past medical history and family history were also unremarkable. Physical examination was normal apart from the right-sided abdominal mass that was firm but non-tender.

Laboratory examinations, including full blood count and extensive biochemical profile, were all within normal limits. Ultrasound examination showed a large cyst of 13.7 cm in maximum diameter in the upper right abdomen. On computed tomography (CT), the cyst was situated in the right retroperitoneum, it was about 13 cm in vertical height, anterior to the right kidney, below the right lobe of the liver and below the gallbladder (Figure 1). It was unilocular and had a homogeneous fluid density. It appeared to be separate from the head of the pancreas. The kidneys and urinary tract appeared normal. There was a small cystic structure just above the uterus, presumably a right ovarian cyst. No other abnormality was detected with respect to the uterus, ovaries or fallopian tubes.

Laparotomy was performed 1 month after clinical presentation. A large cyst was found in the retroperitoneum, situated partially within and behind the mesentery of the hepatic flexure of the colon. There was no attachment to, or shared blood supply with, any abdominal or pelvic organs. The cyst was partially decompressed and then mobilized without difficulty and removed.

Macroscopically, the cyst was unilocular, uniformly thin-walled, and measured 13 cm in maximum diameter. It contained straw-coloured fluid. Histological examination revealed an inner lining of simple ciliated epithelium of Mullerian type, with an outer fibrous layer resembling ovarian stroma. No smooth muscle was present in the wall. There was no stromal or epithelial atypia (Figure 2). Early post-
operative recovery was uneventful and no problems were noted at 3-month follow-up.

Discussion

Retroperitoneal cysts can be categorized on an embryological and histogenetic basis, and include lymphatic cysts, mesothelial cysts, enteric cysts and urogenital cysts. Urogenital cysts can be further subclassified into pronephric, mesonephric, metanephric and Mullerian types.2

The pathogenetic mechanism for the development of Mullerian epithelium-lined cysts in the retroperitoneum is not clear. Three main theories are postulated. Retroperitoneal tissue may include aberrant embryologically-derived Mullerian duct remnants that might have the capacity to grow in later life under the influence of abnormal hormonal stimuli. Alternatively, the coelomic epithelium or peritoneum may undergo differentiation to become serous/tubal-type epithelium, later invaginating into the underlying tissue and eventually losing its connection with the surface, producing a cystic structure.3 Some authors believe that ectopic endometrial tissue (endometriosis), as transplanted by retrograde menstruation or following pelvic surgical instrumentation, may give rise to retroperitoneal cysts.4

Clinically, retroperitoneal cysts appear at any age, but have a peak incidence in the fourth decade of life. The least number of cases are found in patients in their first and sixth decades of life. Most series show a slight female preponderance.2

The cysts may be asymptomatic, as in our case, and appear as an incidental finding of an abdominal mass. Alternatively, they may cause chronic abdominal complaints, the main one being abdominal pain followed by nausea and vomiting. In a minority of cases, mesenteric cysts present as an acute abdomen, due to haemorrhage, rupture or torsion of the cyst, or more rarely with infection within the cyst.5 Hydronephrosis and hydroureret have been described as secondary to ureteral obstruction, and there are a few cases of malignant degeneration of the cysts into low-grade sarcomas.5–7 The reported incidence of malignant forms is less than 3%.8

Figure 1. Computed tomography scan of the abdomen: (A) transverse cross section showing a 12.5 × 7.7 cm, right-sided abdominal cyst; (B) coronal section demonstrating the craniocaudal extent of the cyst.

Figure 2. Haematoxylin and eosin stain showing the cyst with an inner lining of ciliated tubal-type (serous) epithelium and a surrounding zone of fibrous tissue.
The diagnosis of mesenteric cyst is difficult preoperatively due to the lack of pathognomonic clinical features. The classical presentation is an abdominal mass, mobile in the transverse but not the longitudinal plane, although most series show a palpable mass present in only 50% of cases.\(^7\,9\) Laboratory data do not contribute to the diagnosis. The role of radiology is to demonstrate the cystic nature of the abdominal mass, but only with surgery can the definitive histological diagnosis be established. After discovering a cystic abdominal mass, an attempt should be made to determine the organ from which it originates.\(^10\) Ultrasound is the most efficient procedure for detection, although this method’s precision for visceral localization of cysts is not entirely reliable.\(^11\) The cyst’s relationship with the intestinal tract may be evaluated with upper gastrointestinal series, and the adjacent organs to the cyst may be delineated with the help of CT. Magnetic resonance imaging is the best radiological method to determine the nature of the cystic lesion, such as serous or bloody.\(^12,13\)

Optimally, these cysts should be completely excised with minimal disturbance of the surrounding normal structures. In those instances where the bowel is adherent to the cyst wall, a segmental bowel resection should be carried out due to the involvement of mesenteric vessels.\(^9,14,15\) Other treatment options include external marsupialization or internal drainage, both of which are associated with a high rate of morbidity and risk of recurrence. Simple aspiration should be discouraged because recurrence rates are high.\(^5,9\)

**References**